

CASE REPORT

Acute myocardial infarction as a cause of death in palliated hypoplastic left heart syndrome

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A 20 month old child with hypoplastic left heart syndrome died suddenly from a massive myocardial infarction 15 months after a hemi-Fontan operation. This was confirmed at postmortem examination and histological examinations. The sites of surgical reconstruction were all in good condition, there were no gross anatomical coronary abnormalities, and the coronary ostia were unobstructed. On microscopy the internal coronary arteries had notable intimal and medial thickening with narrowing of the lumen, although no thrombotic occlusion was seen. To the authors' knowledge, this is the first published report of arteriosclerosis of the coronary arteries in hypoplastic left heart syndrome. It raises the question as to whether there may be a primary histological abnormality in some children with this condition or whether some mechanism of accelerated arteriosclerosis is at work.

A 20 month old boy with hypoplastic left heart syndrome (HLHS) had a sudden cardiac arrest at home. After antenatal diagnosis with mitral stenosis and aortic atresia, he had successfully undergone a Norwood procedure at two days of age followed by a hemi-Fontan operation at five months. At follow up his growth and development were within normal limits and he was on no medication. His echocardiogram showed good right ventricular function with minimal tricuspid regurgitation and unobstructed flow through the hemi-Fontan anastomosis and aortic arch.

His parents performed cardiac massage, which was continued by paramedics. He was fully resuscitated at his local hospital and a spontaneous cardiac output was restored after 90 minutes. He was transferred to the paediatric intensive care unit at Guy's Hospital, where his ECG showed ST segment changes suggestive of inferolateral infarction. His echocardiogram showed poor right ventricular function, with hypokinesia of the inferolateral wall. Despite maximal inotropic support he sustained another arrest and after an initial attempt at resuscitation, treatment was withdrawn.

At postmortem examination the right ventricular wall was thickened and, with the muscle around the tiny left ventricle, was very pale. Histology showed massive infarction. No gross anatomical coronary abnormalities were found. The ostia were unobstructed but on microscopy the internal coronary arteries had notable intimal and medial thickening with narrowing of the lumen (figs 1 and 2). No thrombotic occlusion was seen. The right sided heart valves appeared normal and the sites of surgical intervention were all in good condition, including the reconstructed arch.

DISCUSSION

Reconstructive surgery for HLHS is undertaken in three stages over three to four years, gradually separating the



Figure 1 Infarcted cardiac muscle around arteriosclerotic intramyocardial coronary artery. Elastic van Gieson stain, low power.

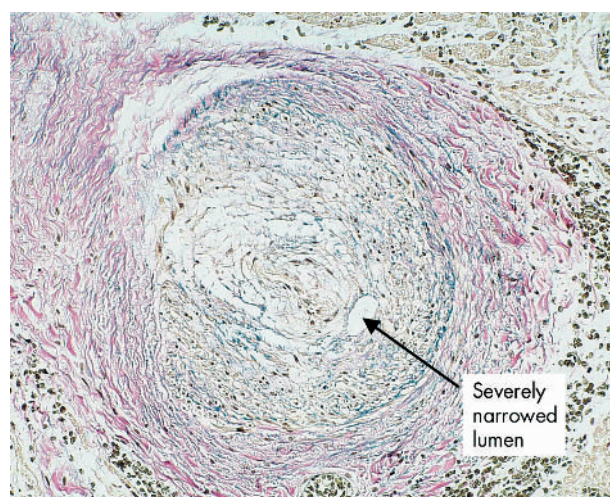


Figure 2 Intimal and medial hyperplasia with elastic reduplication, resulting in a narrowed lumen. Elastic van Gieson stain, high power.

systemic and pulmonary circulations. Ultimately systemic venous blood is directed straight to the pulmonary arteries and the right ventricle becomes exclusively the systemic pump. The mortality after stage I (the Norwood procedure) is relatively high (41% in our recent series¹), but is very much lower (5% or less) after stages II and III (the hemi-Fontan and Fontan operations). At follow up, many survivors have a good quality of life and normal intelligence.²

Although the majority of deaths occur during or shortly after surgery, there seems to be a small but significant risk of sudden death between operations, highest between stages I and II. After stage I, causes of death include residual lesions such as shunt occlusion or aortic arch obstruction, neurological events, and arrhythmias.³ However, between stages II and III or after a modified Fontan procedure, the cause of death is often unclear.

There is increasing evidence that abnormal coronary artery perfusion may contribute to this late attrition. In untreated HLHS, coronary perfusion is largely dependent on retrograde flow through the hypoplastic ascending aorta (or is entirely dependent in cases of aortic atresia). If the degree of hypoplasia is extreme, transient episodes of fetal or neonatal distress may lead to coronary hypoperfusion and result in myocardial ischaemia.^{4,5} Donnelly and colleagues⁶ used positron emission tomography to show that myocardial perfusion was worse in infants who had undergone stage I surgery for HLHS, in which the right ventricle bears the systemic load, than in those undergoing repair of other forms of congenital heart disease. In addition, the pattern of coronary blood flow appears to be abnormal in some infants. In patients with aortic atresia who had undergone stage I, Fogel and colleagues⁷ showed that coronary blood flow occurs primarily in systole, with run off into the aortopulmonary shunt during diastole, limiting endocardial perfusion. However, after the hemi-Fontan procedure, coronary blood flow occurs mainly in diastole, which may be one explanation for the reduced risk of sudden death after stage II.

This report describes a child with HLHS who died of acute myocardial infarction 15 months after stage II, in whom arteriosclerosis of the internal coronary arteries was found at postmortem examination. Although the studies cited above go some way to explaining the risk of sudden death in palliated HLHS, particularly in the months after stage I, to our knowledge this is the first report of sclerosis of the coronary arteries being found at postmortem examination.

The question is raised as to whether there may be a primary histological abnormality of the coronary arteries in some children with HLHS or whether some mechanism of accelerated arteriosclerosis is at work. As our experience of managing patients with palliated HLHS increases, we hope that an improved understanding of the pathophysiological mechanisms responsible for sudden deaths will lead to identification of those at greatest risk and ultimately to a strategy for prevention.

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